BMJ Case Reports

TITLE OF CASE

‘Should have gone to…’: bilateral papilloedema with normal CSF Pressure due to Vestibular Schwannoma

SUMMARY

A 24-year-old woman presented with bilateral blurring of her distance vision and ‘dizzy spells’. She had no other neurological symptoms or past medical history. She attended an optometrist and Optical Coherence Tomography (OCT) was performed which demonstrated papilloedema. She was referred to the local Eye Clinic for assessment and from there was referred for neurological assessment.

Her initial investigations revealed no abnormalities and brain imaging was reported as normal. In the absence of an alternative diagnosis idiopathic intracranial hypertension was considered and a lumbar puncture was performed. This showed elevated protein but normal CSF pressures. MRI the next day revealed a large cerebello-pontine lesion in keeping with vestibular schwannoma. She was referred to Neurosurgery for operative management.

This case highlights three interesting points; the aetiology of her papilloedema without raised intracranial pressure; the decision to perform a lumbar puncture in suspected IIH; and community OCT as a clinical adjunct.

BACKGROUND

Vestibular Schwannoma is a rare, benign primary intracranial tumour of the myelin forming cells of the vestibulcochlear nerve. Common signs and symptoms include; tinnitus, unsteadiness, vertigo and hearing loss. Papilloedema typically manifests late in Vestibular Schwannoma but can be found in 8% of presenting patients.

We report a case of Vestibular Schwannoma with bilateral papilloedema and visual disturbance as the primary presenting feature.

CASE PRESENTATION

A 24-year-old woman presented with six months of bilateral blurring of her distance vision and ‘dizzy spells’ where her vision darkened and resolved spontaneously over seconds. She had no headache, vertigo, hearing loss or other neurological symptoms. There had been no recent weight gain. There was no past medical history of note. She was not taking any regular medication. There was no family history of neurological illness.

She attended an optometrist and Optical Coherence Tomography (OCT) was performed which demonstrated papilloedema. She was referred to the local Eye Clinic and from there for neurological assessment.

Visual acuity was 6/5 bilaterally. Visual fields were normal to confrontation. External ocular movements and pupillary responses were normal with bilateral papilloedema on fundoscopy. There was no loss of facial sensation, hearing, facial weakness, dysarthria or dysphagia. Peripheral nervous system examination was normal. Her body-mass index was 33kg/m^2. Blood pressure was 110/70.

Non-contrast CT Brain was reported as showing no evidence of a space occupying lesion(SOL). There was no evident sinus thrombosis. Ventricular size was normal. This was interpreted

clinically as supporting a diagnosis of Idiopathic Intracranial Hypertension (IIH).
MRI and MR Venogram of Brain were requested but not immediately available. Lumbar Puncture(LP) was performed and showed opening pressure of 21cm H_2O (6-25cm H_2O) with raised CSF protein; 1.34 g/L(0.-0.4g/L).
MRI Brain the next day demonstrated a 2.6cm extra-axial right-sided cerebello-pontine angle lesion that enhanced post contrast. The right internal auditory meatus was asymmetrically widened favouring the characteristic “ice-cream cone” appearance of Vestibular Schwannoma (figure 1). There were no features of hydrocephalus or malignancy. MR Venogram was normal.
Retrospectively the initial CT was abnormal. The pons was displaced to the left with an associated mass in the right cerebello-pontine angle (figure 2).
She was referred to neurosurgery – Vestibular Schwannoma was confirmed postoperatively.

OUTCOME AND FOLLOW-UP

Post-operatively the patient is doing well. She reports some mild unilateral hearing loss which is persistent but improving. Her visual deterioration and dizzy spells have resolved completely and she is under follow-up with both the Neurological and Neurosurgical teams. Post-operative and interval imaging have been stable.

DISCUSSION *Include a very brief review of similar published cases*

This case illustrates useful teaching points. The initial impression was IIH, despite the absence of headache, given its prevalence in younger women. This was supported by the CT report. The lesion would likely have been detected on contrast enhanced CT had it been performed. This raises the suggestion that either contrast enhanced CT or MRI be performed before LP in suspected IIH.
Papilloedema typically manifests late in Vestibular Schwannoma but can be found in 8% of presenting patients. This has been ascribed to raised ICP secondary to obstructive hydrocephalus or mass effect. In this case the pressure was normal. Case reports of papilloedema without evidence of raised ICP propose possible mechanisms including: CSF protein secretion from the tumour causing impaired CSF reabsorption and a ‘communicating’ hydrocephalus - this mechanism has been previously hypothesised in association with spinal cord schwannomas[1]; or early hydrocephalus not yet manifesting on CT; or that the pressure is raised in a pulsatile fashion[2]. In our case there was significant displacement of the brainstem. This may have led to reduced CSF flow and lower pressure below the lesion.
Lastly this case highlights the role of community opticians in detecting eye and intracranial pathology. Many opticians now have access to OCT - a reliable tool for the diagnosis of papilloedema in young adults[3].

LEARNING POINTS/TAKE HOME MESSAGES *3-5 bullet points*

1. *Atypical disease presentations;* Though not a typical presenting complaint, in patients presenting with papilloedema and normal intracranial pressure Vestibular Schwannoma should be considered.

2. *Imaging modalities;* Had the patient had a contrast enhanced CT Brain it is likely that the lesion would have been seen with more prompt diagnosis and avoidance of invasive procedures. In similar presenting cases either contrast enhanced CT or MRI should be considered, particularly prior to invasive procedures.

3. *Community diagnostics;* With the advent of community OCT we see an increasing role for community opticians in the detection of eye and intracranial pathology.

REFERENCES

References:

1. Feldmann E, Bromfield E, Navia B, Pasternak GW, Posner JB. Hydrocephalic dementia and spinal cord tumor. Report of a case and review of the literature. Arch Neurol. 1986 Jul;43(7):714–8.
2. Matos RJC, Gil PNBCQ, Pires JMS, Lopes N. Advanced Vestibular Schwannoma: A Case of Optic Disc Oedema without Hydrocephalus. Neuroophthalmology. 2016 Oct;40(5):222–4.
3. Martinez MR, Ophir A. Optical coherence tomography as an adjunctive tool for diagnosing papilledema in young patients. J Pediatr Ophthalmol Strabismus. 2011 Jun;48(3):174–81.

FIGURE/VIDEO CAPTIONS

Figure 1: *Axial MRI Brain*, *T2:* A 2.6cm extra-axial mass can be seen in the right CPA with associated asymmetrical widening of the right internal auditory meatus. This feature is commonly described as the “ice-cream cone sign” or “ice-cream cone” appearance.

Figure 2: *Axial Non-contrast enhanced CT Brain:* Effacement of the pons to the left can be seen(blue arrow) with an associated mass in the right cerebello-pontine angle(grey arrow).

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